

# Kawasaki Disease in Children

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## Abstract

Kawasaki Disease (KD), previously defined as mucocutaneous lymph node syndrome, is a febrile illness caused by inflammation of the body's medium-sized blood vessels. Tomisaku Kawasaki first described it in 1967 as a self-limited acute vasculitic syndrome of unknown etiology. Intravenous immunoglobulin (IV) and high-dose oral aspirin should be begun within 10 days of disease onset along with aspirin.

**Keywords:** Kawasaki disease; Fever; Coronary artery; Immunoglobulin

**Abbreviations:** KD: Kawasaki Disease, ESR: Erythrocyte Sedimentation Rate, CRP: C-reactive protein, MIS-C: Multisystem Inflammatory Syndrome in Children, NT-proBNP: Nterminal-prohormone brain natriuretic peptide

## Introduction

Kawasaki Disease (KD), previously defined as mucocutaneous lymph node syndrome, is a febrile illness caused by inflammation of the body's medium-sized blood vessels. Tomisaku Kawasaki first described it in 1967 as a self-limited acute vasculitic syndrome of unknown etiology. KD has also passed rheumatic fever as the leading cause of acquired heart disease in young children in many countries. More than 80% of cases include young children under the age of five. It affects boys more than girls (M: F 1.5:1). While the exact cause of KD remains unclear, many hypotheses exist, including an infectious disorder triggered by bacteria or bacterial superantigens (streptococcus pyogenes) or a virus infectious etiology, hereditary makeup, and an immunological abnormality. There is no clear lab procedure that can be used to definitively diagnose KD. KD is diagnosed depending on the appearance of specific clinical symptoms.

Classic KD needs the presence of fever for at least 5 days and at least four of five of the other signature clinical characteristics of illness. In atypical KD, the patient has a persistent fever but only one or two other symptoms of the disease [1-2].

**The diagnostic criteria for classic KD are as follow:**

- a. Fever for  $\geq 5$  days
- b. Presence of  $\geq 4$  principal features:
  - i. Erythema, swelling, and desquamation of the extremities. Fingernail and toe desquamation
  - ii. A polymorphous (non-vesicular) rash is usually generalized, although it may be limited to the genital or lower extremities.
  - iii. Bilateral bulbar conjunctival injection without exudate



- iv. Lip and oral cavity: erythema, lip splitting, strawberry tongue, and diffuse injection of oral and pharyngeal mucosae are all symptoms.
- v. Cervical lymphadenopathy (diameter greater than 1.5 cm), typically unilateral
- c. There is no other recognized disease mechanism that may justify a symptom.

The above-mentioned clinical characteristics appear sequentially over a few days and do not all have to be present at the same time. Other clinical manifestations of the condition may include aseptic meningitis, urethritis, orchitis, arthritis/arthralgia, stomach pain, vomiting, diarrhea, sterile pyuria, hepatitis, and gallbladder distention. Myocarditis, pericarditis, pericardial effusion, or reduced ventricular activities are also examples of cardiac involvement. Around 25% of untreated patients have coronary artery aneurysms in the second to third week of illness [2-3].

### **KD is generally divided into three clinical phases**

- **The Acute febrile phase**, which typically lasts 1–2 weeks and is characterized by fever and other acute illness symptoms. On the hands and feet, erythema and edema appear. The tongue and oral mucosa turn bright red and begin to crack. Myocarditis and pericarditis are also cardiac conditions.
- **The Subacute phase**, Irritability, anorexia, and conjunctival injection may occur after the fever and other acute symptoms have subsided. This procedure should be finished by the fourth week. This stage is associated with desquamation, thrombocytosis, and the development of coronary aneurysms. Children are most vulnerable to sudden death at this time.
- **The Convalescent phase**, begins 6–8 weeks after the onset of disease and continues until all clinical signs have resolved and the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) have started to improve. The presence of coronary artery aneurysms is a significant clinical finding that persists throughout this time span.

### **Investigation**

- Leukocytosis accompanied by neutrophilia and immature types
- Normocytic, normochromic anemia
- Raised ESR, CRP, and other acute-phase reactants are almost universally present.
- The platelet count is typically average in the first week of illness and rapidly increases by the second–third week, often exceeding 1,000,000/ mm<sup>3</sup>.
- Hyperlipidemia (abnormal plasma lipids)
- Low albumin levels
- Rheumatoid factor and antinuclear antibody tests are negative.
- Sterile pyuria, minor hepatic transaminase elevations, and cerebrospinal fluid pleocytosis can be present.
- Serum transaminases and gamma-glutamyl transpeptidase levels are elevated.
- Two-dimensional echocardiography: Monitoring the occurrence of coronary artery aneurysms [3-5].

### **Differential Diagnosis**

Measles, Adenovirus pneumonia, scarlet fever, Toxic shock syndrome, Drug hypersensitivities including Stevens-Johnson syndrome, Leptospirosis, Juvenile rheumatoid arthritis, and MIS-C (Multisystem Inflammatory Syndrome in Children).

**The complication of KD** is aneurysms of the coronary arteries and rupture. These aneurysms will lead to heart attacks later in life. Dehydration and reduced movement due to joint inflammation are two other risks.

### **Treatment for KD**

Intravenous immunoglobulin (IV) and high-dose oral aspirin should be begun within 10 days of disease onset.

**Acute stage:** 2 g/kg IV immunoglobulin given over 10-12 hours, with aspirin 80-100 mg/kg given orally every 6 hours before the 14th illness day.

**Convalescent stage:** For 6-8 weeks after the start of the infection, take 3-5 mg/kg aspirin orally once daily. Long-term therapy for coronary heart disease involves aspirin 3-5 mg/kg once daily and clopidogrel 1 mg/kg once (max 75 mg/day).



Patients with large or multiple aneurysms can require clopidogrel, warfarin, or low molecular weight heparin treatment [4-5].

Multisystem Inflammatory Syndrome in Children is a significant differential diagnosis of KDs [6]. The WHO category of the MIS-C applies to children and adolescents aged 0 to 19 years who follow all of the clinical criteria mentioned below:

- Febrile illness for  $\geq 3$  days
- 2 or more of the following:
  - Hypotension or shock.
  - Increased levels of nonspecific inflammatory markers (e.g., erythrocyte sedimentation rate, C- reactive protein, procalcitonin).
  - Severe gastrointestinal complaints (vomiting, diarrhea) or stomach pain There are no clear alternative microbial causes of inflammation (bacterial sepsis, staphylococcal, or streptococcal toxic shock syndrome)
  - Characteristics of myocardial dysfunction, pericarditis, valvulitis, or coronary artery defects, including proof discovered by imaging (echography) and experimental tests (elevated levels of troponin, NT-proBNP [Nterminal-prohormone brain natriuretic peptide]).
  - Rash or non-purulent bilateral conjunctivitis, or mucocutaneous inflammation of the lips, hands, or feet.
  - Coagulation profile (for example, elevated prothrombin time/INR, partial thromboplastin time, D-dimer level)
- COVID-19 evidence (positive reverse transcription-polymerase chain reaction test response, detectable antigen or antibody) or probable COVID-19 exposure

Some children follow complete or partial conditions for Kawasaki syndrome, but they are diagnosed with MIS-C if they otherwise match the case description.

### **Certain characteristics of MIS-C are shared but not typical for classic Kawasaki disease:**

- Abdominal pain is a normal symptom, and its severity is comparable to that of classic Kawasaki disease.
- MIS-C signs include thrombocytopenia, anemia, and lymphopenia.
- The levels of ferritin, troponin, proBNP, and D-dimer are all increased.

The treatment of the MIS-C is the used of IV immunoglobulin in a single dose of 1-2 gm /kg IV in combination with aspirin with steroid therapy.

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